Agency for Healthcare Research and Quality
Advancing Excellence in Health Care

NATIONAL QUALITY MEASURES CLEARINGHOUSE

General

Title

Amyotrophic lateral sclerosis (ALS): percentage of patients with a diagnosis of ALS with whom the clinician discussed disease modifying pharmacotherapy (riluzole) to slow ALS disease progression at least once annually.

Source(s)

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

Measure Domain

Primary Measure Domain

Clinical Quality Measures: Process

Secondary Measure Domain

Does not apply to this measure

Brief Abstract

Description

This measure is used to assess the percentage of patients with a diagnosis of amyotrophic lateral sclerosis (ALS) with whom the clinician discussed modifying pharmacotherapy (riluzole) to slow ALS disease progression at least once annually.

Rationale

Riluzole is approved by the Food and Drug Administration (FDA) for slowing disease progress in amyotrophic lateral sclerosis (ALS), and it is the only currently available disease modifying pharmacotherapy for ALS. Riluzole was the subject of a practice advisory published by the American Academy of Neurology (AAN) in 1997 (AAN, Quality Standards Subcommittee, 1997). The practice advisory recommended riluzole 50 mg twice a day (BID) to prolong survival for those with definite or probable ALS less than 5 years duration, with forced vital capacity (FVC) greater than 60%, and without tracheostomy.

Expert opinion suggested potential benefit for those with suspected or possible ALS with symptoms longer than 5 years, FVC greater than 60%, and tracheostomy for prevention of aspiration only. Since 1997, 2 other controlled clinical trials have been published (Bensimon et al., 2002; Yanagisawa et al., 1997) and all of the available evidence has been reviewed (Miller et al., 2007). Riluzole has a modest beneficial effect in slowing disease progression (prolonged survival of 2 to 3 months) based on 4 Class I trials. The number needed to treat to delay 1 death until after 12 months was 11. However, 5 studies using large databases spanning 5 to 10 years have suggested that treatment with riluzole might be associated with a prolonged survival of 6 months (Meininger et al., 2004), 10 months (Mitchell, O'Brien, & Joshi, 2006), 12 months (Traynor et al., 2001), 14 months (Brooks et al., 2001), or even 21 months (Turner et al., 2001). These cohort studies had longer-term follow-up than the clinical trials, but are subject to greater bias. After 10 years of patient experience, the drug appears to be safe but expensive. In fact, the cost does limit access to the drug for a significant portion of patients (Miller et al., 2007; Bryan et al., 1997). Fatigue and nausea are known side effects. Riluzole is safe and effective for slowing disease progression to a modest degree in ALS.

The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:

> Riluzole should be offered to slow disease progression in patients with ALS (Miller et al., "Drug, nutritional," 2009).
> Riluzole 50 mg twice a day is reasonably safe and probably prolongs median survival by about two to three months in patients with ALS (Miller et al., "Riluzole," 2009).
> ALS patients should be offered treatment with riluzole 50 mg twice daily (Andersen et al., 2005).
> Patients treated with riluzole should be monitored regularly for safety (Andersen et al., 2005).
> Treatment should be initiated as early as possible after the patient has been informed of the diagnosis taking into account expected therapeutic benefits and potential safety issues. Realistic expectations for treatment effects and potential side effects should be discussed with the patient and caregivers (Andersen et al., 2005).
> Treatment with riluzole should be considered in progressive muscular atrophy (PMA) and primary lateral sclerosis (PLS) patients who have a first degree relative with ALS (Andersen et al., 2005).

Evidence for Rationale

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Primary Health Components

Amyotrophic lateral sclerosis (ALS); disease modifying pharmacotherapy; riluzole

Denominator Description

All patients with a diagnosis of amyotrophic lateral sclerosis (ALS) (see the related "Denominator Inclusions/Exclusions" field)

Numerator Description

Patients with whom the clinician discussed disease-modifying pharmacotherapy (riluzole) to slow amyotrophic lateral sclerosis (ALS) disease progression at least once annually

Evidence Supporting the Measure

Type of Evidence Supporting the Criterion of Quality for the Measure

Type of Evidence Supporting the Criterion of Quality for the Measure

A clinical practice guideline or other peer-reviewed synthesis of the clinical research evidence

A formal consensus procedure, involving experts in relevant clinical, methodological, public health and organizational sciences

A systematic review of the clinical research literature (e.g., Cochrane Review)

One or more research studies published in a National Library of Medicine (NLM) indexed, peer-reviewed journal

Additional Information Supporting Need for the Measure

Importance of Topic

Prevalence and Incidence

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, is a type of motor neuron disease that is a rapidly progressive and fatal neurological disease (National Institute of Neurological Disorders and Stroke [NINDS], 2013).
Twenty thousand to 30,000 people in the United States (U.S.) have ALS (NINDS, 2013).
Five thousand people are diagnosed with ALS in the U.S. annually (NINDS, 2013).
ALS is one of the most common neuromuscular diseases worldwide (NINDS, 2013).
In 90% to 95% of all ALS cases the disease occurs apparently at random with no clearly associated risk factors (NINDS, 2013).
Five percent to 10% of all ALS cases are inherited (NINDS, 2013).
Twenty percent of all familial cases result from a specific genetic defect that leads to mutation of the enzyme known as superoxide dismutase 1 (SOD1) (NINDS, 2013).
No cure exists for ALS. Newer pharmacotherapy agents have been found to reduce the progression, but not halt the disease development (NINDS, 2013).
The prevalence of ALS is said to be between six and eight cases per 100,000 in the population. Using the higher prevalence estimate and data from the 2000 U.S. census, nearly 22,600 Americans are living with ALS at any one time. Since ALS is a disease of aging, as the U.S. population increases and ages, an increase in the prevalence of ALS can be anticipated (ALS Association, 2012)
Cognitive dysfunction is seen in 20% to 50%, while only 3% to 5% develop dementia that is usually of frontotemporal type (Strong et al., 2009). Consensus criteria for diagnosis have recently been reported (Strong et al., 2009).
Death due to respiratory failure follows on average 2 to 4 years after onset, but a small group may survive for a decade or more (Haverkamp, Appel, & Appel, 1995).
The mean age of onset is 47 to 52 years in familial cases (FALS) and 58 to 63 years in sporadic (SALS) cases (Bobowick & Brody, 1973).
The lifetime risk for developing ALS for individuals aged 18 years has been estimated to be 1 in 350 for men and 1 in 420 for women (Armon, 2007) with male sex, increasing age and hereditary disposition being the main risk factors (Heffernan et al., 2006).

Mortality and Morbidity

Most patients with ALS die within 2 to 5 years of onset (Lechtzin et al., 2002). Only 10% of ALS patients survive for 10 years or more (Miller et al., "Drug, nutritional," 2009).
Treatment of respiratory insufficiency improves survival, quality of life and respiratory symptoms (Lechtzin et al., 2002; Miller et al., "Drug, nutritional," 2009). The diagnosis and management of respiratory insufficiency is critical because most deaths from ALS are due to respiratory failure (Lechtzin et al., 2002; Miller et al., "Drug, nutritional," 2009; EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis et al., 2012; Laird et al., 2001).
Falls surveillance will lead to interventions to prevent falls and decrease fall related deaths in ALS patients. Falls are an independent predictor of adverse health outcomes (Gil et al., 2008). Fall

related deaths occur in 1.7% of ALS patients (Rubenstein & Josephson, 2002). Several specific risk factors for falls have been identified, including muscle weakness, deficits in gait or balance, visual deficits, arthritis, impairments in activities of daily living, depression, and cognitive impairment (Ringholz et al., 2005).

Studies confirm the presence of cognitive impairment in 50% of patients with ALS and particularly implicate executive dysfunction and mild memory decline in the disease process (Laird et al., 2001). More severe impairment occurs in a subset of patients with ALS and has features consistent with frontal temporal dementia (FTD) (Phukan, Pender, & Hardiman, 2007; Gordon et al., 2007). Recent studies have demonstrated the feasibility of screening patients in a busy specialized ALS clinic (Flaherty-Craig et al., 2009; Woolley & Katz, 2011), but this is still not routinely practiced. A fuller characterization of the extent of cognitive and behavioral dysfunction in ALS has important implications given that it shortens survival (Elamin et al., 2011), and the burden and stress for carers of patients with FTD is very great. It also has relevance to effective communication, legal issues and end-of-life decision making by patients with motor neuron disease (MND) (Elamin et al., 2011).

Pseudobulbar affect (PBA), excessive laughing or crying, or involuntary emotional expression disorder affects 20% to 50% of patients with ALS, especially in pseudobulbar palsy (McCullagh et al., 1999). Patients are embarrassed and isolated by these symptoms, which in turn greatly diminishes the patients' quality of life.

Sialorrhea, or drooling, is embarrassing, socially isolating, and is associated with aspiration pneumonia. The prevalence is estimated at 50%, and 70% of patients receiving oral medications for treatment reported benefit (Laird et al., 2001; Miller et al., "Multi-disciplinary," 2009)

Fatigue may be a symptom of depression, poor sleep, abnormal muscle activation, immobility, or respiratory dysfunction. Fatigue diminishes quality of life for patients with ALS. Fatigue was a side effect of therapy in 26% of patients taking riluzole vs. 13% taking placebo (Bensimon, Lacomblez, & Meininger, 1994). Asthenia occurred in 18% of patients taking riluzole vs. 12% of patients taking placebo in a larger study (Lacomblez et al., 1996).

The prevalence of depression in ALS ranges from 0% to 44%, although systematic studies suggest 10% in advanced ALS (Laird et al., 2001; Wicks et al., 2007). Depression shortens survival and lowers quality of life for patients with ALS (Phukan, Pender, & Hardiman, 2007). There is consensus among experts that depression should be treated in patients with ALS (Laird et al., 2001); however, there are no controlled studies of benefit or harm.

Insomnia is common in ALS and may be a symptom of early respiratory weakness, underlying anxiety, depression, or pain (Hetta & Jansson, 1997). There is a concern that sedative/hypnotic agents may suppress the respiratory drive in patients with ALS.

Weight loss is a key prognostic indicator for ALS with the risk of death increased 7-fold when body mass index is less than 18.5 kg/m^2 (Marin et al., 2011; Lehéricey et al., 2012; Spataro et al., 2011; Desport et al., 1999; Vaisman et al., 2009; Dupuis et al., 2008).

ALS patients have dysarthria in nearly all bulbar onset patients and nearly 40% of ALS patients with spinal onset. More than 95% of ALS patients cannot speak before death and patients who accept gastrostomy tube, non-invasive ventilation or tracheostomy-ventilation have a greater need for augmentative alternative communication as the disease progresses (Ball, Beukelman, & Pattee, "Communication," 2004; Ball, Beukelman, & Pattee, "Acceptance," 2004; Mathy, Yorkston, & Gutmann, 2000; Beukelman, Fager, & Nordness, 2011).

End of life discussions will improve patient decision making with respect to disease management (NINDS, 2013; ALS Association, 2012; Strong et al., 2009; Haverkamp, Appel, & Appel, 1995; Bobowick & Brody, 1973; Heffernan et al., 2006). Pain in ALS should be treated following accepted guidelines (Oliver et al., 2011; Albert et al., 1999, Mitsumoto et al., 2005; Nolan et al., 2008; Albert et al., 2005; Albert et al., 2009).

Office Visits and Hospital Stays

One study's significant findings were that common morbidities increased over time (pneumonia [38.1% to 47.3%], respiratory failure [26.9% to 35.5%], and nutritional deficiency [43.0% to 56.3%]); the median length of stay dropped from 6 to 4 days; mean hospital charges increased from

$21,574 to $24,314; the proportion of hospital deaths decreased over time (17.6% to 14.6%), whereas the proportion discharged to home health/hospice care (14.0% to 18.2%) and to long-term care facilities (13.2% to 27.9%) increased. The odds ratio (OR) of death was 5.03 (95% CI: 4.57 to 5.54) for those admitted with respiratory failure, 1.36 (1.24 to 1.50) for those with pneumonia, and 0.84 (0.77 to 0.92) for those with nutritional deficiency. The high OR of death in patients admitted for pneumonia or respiratory failure is likely associated with more advanced disease, whereas the protective effect of admission for nutritional deficiency is consistent with the predominance of bulbar symptoms and admission earlier in the disease. The trends during the 15 years of this administrative data set were for increasing comorbidities and higher utilization of end-of-life care (Dubinsky, Chen, & Lai, 2006).

Family Caregiving

Caregiver burden was correlated to their level of depression and quality of life and, differently from other chronic disorders, increased with the worsening of patients' disability. ALS patients have a good objective perception of their impact on caregivers (Chiò et al., 2005).
Recent studies assessing caregivers' burden in chronic neurologic disorders have found some features shared by caregivers: the perceived burden exceeds the objective measures of patients' impairment, the amount of burden is independent of diagnosis, and the patients' cognitive functioning is an important factor in determining the level of burden (Thommessen et al., 2002).

Cost

ALS is a difficult to diagnose, fatal, progressive degenerative disease with an average survival time of 2 to 5 years. Percutaneous endoscopic gastrostomy (PEG) and bi-level intermittent positive pressure (BIPAP) ventilation may be the major interventions leading to longer survival of patients with ALS. Riluzole has been shown to have modest effects on survival (as opposed to functional) gains and is currently the only drug approved for the treatment of ALS. Mechanical ventilation (via a tracheostomy tube) is expensive, but is widely used in later stage patients with ALS in the U.S. A review of nine cost-effectiveness studies of riluzole found the following: drug costs and survival gains are the major drivers of cost effectiveness; survival gains are estimated from truncated databases with a high degree of uncertainty; more accurate stage-specific utility weights based on patients who agreed to treatment are needed; case incidence-based evaluations should be carried out; cost-effectiveness ratios are insensitive to discount rates; employment and caregiver issues or externalities have been widely ignored; threshold acceptance cost-effectiveness values are ill-defined and evaluations are not generalizable to other countries because of cost and treatment style differences. On account of the high degree of uncertainty pertaining to survival gains and the relatively high costs per life years or quality-adjusted life-years gained, and while acknowledging that not every therapy has to be cost effective (e.g., orphan drugs), it is still inconclusive as to whether or not riluzole can be considered as cost-effective therapy for ALS (Ginsberg & Lowe, 2002).

Disparities

All races and ethnic backgrounds are affected by ALS (NINDS, 2013).
ALS most common in individuals 40 to 60 years old, but younger and older people can develop the disease (NINDS, 2013).
Men are more likely to develop ALS than women. Studies suggest an overall ratio of about 1.5 men to every woman who develops ALS in Western countries (ALS Association, 2012).

Opportunity for Improvement

Riluzole is currently the only available disease modifying pharmacotherapy available to slow down progression of ALS. Only 60% of patients are taking the riluzole in the U.S., compared to nearly 100% in European countries (France, Italy, Germany) (Miller et al., "Outcomes," 2009). This utilization is improved compared to 45% in 1997, a rise that reflects increased awareness and experience of treating physicians (Bradley et al., 2004). The cost is still a major factor for many patients. These data reflect the utilization of riluzole in large multidisciplinary clinics, and it is much

lower in community-treated patients. Considerable misunderstanding exists around safety and efficacy, both for patients and physicians. More education is needed. The most influential factor in whether patients take riluzole is the knowledge and enthusiasm of the treating physician (Miller et al., "Riluzole," 2009; Bryan et al., 1997). ALS experts in a multidisciplinary clinic are most likely to adequately inform patients about this neuroprotective medication. Also, the more recent registry studies suggesting a much greater survival benefit have been impressive (Miller et al., "Riluzole," 2009).

Evidence for Additional Information Supporting Need for the Measure

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Extent of Measure Testing

This measure is being made available without any prior testing. The American Academy of Neurology (AAN) recognizes the importance of testing of all of its measures and encourages testing of the amyotrophic lateral sclerosis (ALS) measurement set for feasibility and reliability by organizations or individuals positioned to do so. The AAN welcomes the opportunity to promote the initial testing of these measures and to ensure that any results available from testing are used to refine the measures before implementation.

Evidence for Extent of Measure Testing

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

State of Use of the Measure

State of Use

Current routine use

Current Use

not defined yet

Application of the Measure in its Current Use

Measurement Setting

Ambulatory/Office-based Care

Home Care

Hospital Outpatient

Skilled Nursing Facilities/Nursing Homes

Professionals Involved in Delivery of Health Services

not defined yet

Least Aggregated Level of Services Delivery Addressed

Individual Clinicians or Public Health Professionals

Statement of Acceptable Minimum Sample Size

Does not apply to this measure

Target Population Age

Unspecified

Target Population Gender

Either male or female

National Strategy for Quality Improvement in Health Care

National Quality Strategy Aim

Better Care

National Quality Strategy Priority

Person- and Family-centered Care
Prevention and Treatment of Leading Causes of Mortality

Institute of Medicine (IOM) National Health Care Quality Report Categories

IOM Care Need

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Data Collection for the Measure

Case Finding Period

Unspecified

Denominator Sampling Frame

Patients associated with provider

Denominator (Index) Event or Characteristic

Clinical Condition

Denominator Time Window

not defined yet

Denominator Inclusions/Exclusions

Inclusions
All patients with a diagnosis of amyotrophic lateral sclerosis (ALS)

Note: Refer to the original measure documentation for International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) diagnosis codes and Current Procedural Terminology (CPT) Evaluation and Management (E/M) service codes.

Exclusions
None

Exclusions/Exceptions

not defined yet

Numerator Inclusions/Exclusions

Inclusions
Patients with whom the clinician discussed disease-modifying pharmacotherapy (riluzole) to slow amyotrophic lateral sclerosis (ALS) disease progression at least once annually

Exclusions
Unspecified

Numerator Search Strategy

Fixed time period or point in time

Data Source

Administrative clinical data

Electronic health/medical record

Paper medical record

Type of Health State

Does not apply to this measure

Instruments Used and/or Associated with the Measure

Unspecified

Computation of the Measure

Measure Specifies Disaggregation

Does not apply to this measure

Scoring

Rate/Proportion

Interpretation of Score

Desired value is a higher score

Allowance for Patient or Population Factors

not defined yet

Standard of Comparison

not defined yet

Identifying Information

Original Title

Measure #2: disease modifying pharmacotherapy for ALS discussed.

Measure Collection Name

Amyotrophic Lateral Sclerosis Performance Measurement Set

Submitter

American Academy of Neurology - Medical Specialty Society

Developer

American Academy of Neurology - Medical Specialty Society

Funding Source(s)

Unspecified

Composition of the Group that Developed the Measure

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Financial Disclosures/Other Potential Conflicts of Interest

Unspecified

Adaptation

This measure was not adapted from another source.

Date of Most Current Version in NQMC

2012 Jul

Measure Maintenance

Unspecified

Date of Next Anticipated Revision

Unspecified

Measure Status

This is the current release of the measure.

Measure Availability

Source available from the American Academy of Neurology (AAN) Web site .

For more information, contact AAN at 201 Chicago Avenue, Minneapolis, MN 55415; Phone: 800-879-1960; Fax: 612-454-2746; Web site: www.aan.com .

NQMC Status

This NQMC summary was completed by ECRI Institute on March 8, 2016. The information was not verified by the measure developer.

Copyright Statement

Production

Source(s)

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

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